THE CLINICAL FEATURES OF EPIDEMIC ENCEPHALITIS (ENCEPHALITIS LETHARGICA) IN CHILDREN.

by

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Thesis for the Degree of M.D. March 1922.

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Since the commencement of the present epidemic of Encephalitis Lethargica, in 1918, a very large number of papers have been published about the disease as it affects adults, but comparatively little has been written, and that for the most part quite recently, about the disease and its results when it occurs in children.

One of the first references to the disease appears in a paper published by Gordon (1) in 1913, recording four cases, which he examined post mortem. All these cases had occurred within a month, and all were fatal. Three boys and one girl were affected.

In three cases the onset of the illness was marked by drowsiness, one case showed coma, and one delirium. No abnormality was observed in the eyes except "staring". Rigidity of the muscles and twitchings were noticed in all the cases. Abnormal reflexes were also noted. The Cerebro-spinal fluid showed an increase of cells in every case, but no other abnormal constituent.

Post Mortem - the cases showed little except slight

inflammation/

inflammation of the meninges, and cerebral cortex, and inflammation of the parotid.

Gordon suggests that the disease was an abnormal form of Mumps, but in the light of the recent epidemic of encephalitis it is possible that these were cases of the latter disease.

No further contribution on the subject appeared until Batten and Still (2) in 1918, described four cases in children varying in age from 11 - 3½ years.

All these children were lethargic, and slept for 3 - 5 weeks after admission to hospital. They suffered from muscular rigidity, and rhythmic tremor of hands and arms. They had "mask like" expressions. They could be roused with difficulty to swallow food, etc. The cerebro-spinal fluid contained no abnormal constituents and was not under pressure.

After some weeks they wakened up, but suffered from various results, such as a shuffling gait in one, and spastic diplegia in another. Their intelligence was not affected. Nutrition was slightly impaired.

In the same year Finlay (3) described three acute cases of the disease in children, with the onset of lethargy, ocular symptoms, convulsions, vomiting and abnormal reflexes. One case recovered after 5 weeks, one died, and one apparently recovered after/

after 12 weeks, but showed mental deterioration later.

Up to 1920 the somewhat scanty descriptions of the disease as it occurred in children corresponded fairly well with the adult type. In that year however, several independent observers recognized a type of case which was new, and in which the leading symptoms differed markedly from the earlier cases, inasmuch as intractable insomnia, mental excitement and choreic movements occupied a prominent place.

Cases of this kind were observed almost simultaneously in the United Kingdom, United States of America, Germany and Italy. (4)

Among the principal papers dealing with this may be noted those of Happ & Blackfan (5), Hofstadt(6), Finlay & Shiskin (7), Leahy & Sands (8), Happ & Mason (9), Fletcher & Rolleston (19), and Coburn (11).

In these papers the authors describe a series of cases which demonstrate the above-mentioned train of symptoms. In some cases nocturnal insomnia, and diurnal somnolence appeared as a primary symptom; in others it followed preliminary lethargy.

Mental changes, such as defects of character and morals, backwardness, and marked mental defect, are pointed out in a number of these cases, and also by Paterson and Spence. (12) These observers record 25 cases, of which 18 showed definite mental changes at/

at varying periods after an attack of the disease.

In their valuable contribution on the subject, Happ and Mason (9) described 24 cases occurring in children. The ages varied from 4 months to 16 years: 12 were male, 12 female. In 15 cases there was a lethargic onset, followed in four cases by insomnia. In six cases the disease began with intractable sleeplessness. Ocular symptoms were observed in 9 cases, paralysis and abnormal reflexes in 13. Choreiform movements were noted in 7 cases. Mental changes occurred in 5. An onset involving fever occurred in 10 cases. Convulsions accompanied it in 7, delirium in 4, and headache in 9 cases. Of the 24 cases, 13 recovered in periods varying from 10 days to 8 months, six improved, one died, and five remained in statu quo.

Finlay and Shiskin (7) mention 24 cases, with similar symptoms. The ages vary from 2 months to 14 years. 19 were boys and 5 girls.

The onset was lethargic in 18 cases, followed by insomnia in 12 children. Four showed insomnia at the beginning of their illness. Eye symptoms appeared in 11 cases, paralysis and abnormal reflexes in 11, and choreic movements in 17. Mental changes manifested themselves in four cases. At the onset

fever/

fever was present in 11 cases, convulsions in one case only, delirium in six cases, and headache in three. Results were as follows:- 5 recovered completely, 4 showed improvement, 3 died, and 12 remained *in statu quo*.

The following seven cases demonstrate most of the symptoms described by the various authors, and also illustrate the different types of the disease. As none of them throw any light on the pathology it is not proposed to discuss it here.

CASE I. P.L., male, aged 9.

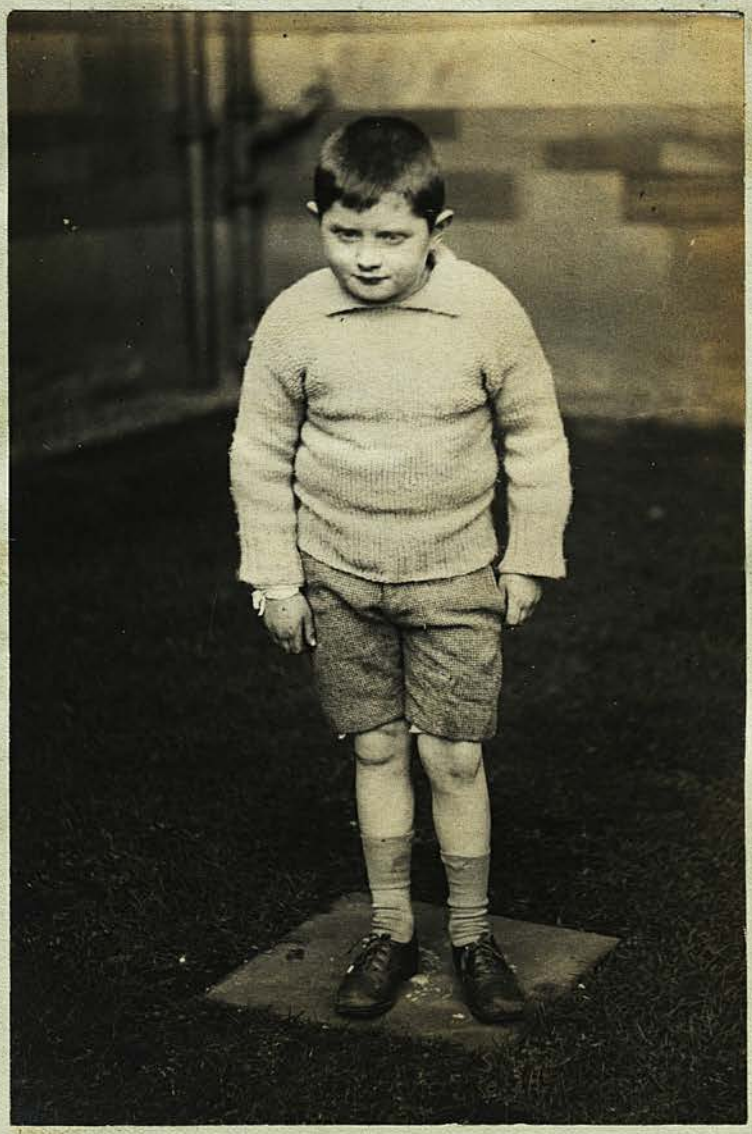
History of Present Illness.

In June 1920 the boy began to suffer from jerking movements of the right foot; these came on quite suddenly one morning, without any previous warning. He very soon became restless at night time, and slept badly, and the irregular movements of his legs increased in severity. Up to the onset of these symptoms he was quite a normal child in every respect - quiet, tractable, and well behaved. The nocturnal restlessness became worse during August and September. He appeared weak and exhausted during the day, but about 8 p.m. he began to get lively and spent the night tossing about and "making his bed". He was never delirious during this period, but was unusually alert mentally, and very talkative; he would get up from bed at night and sit on the window ledge, talking incessantly about the harbour lights and what was going on in the street. Towards the end of September he slept more during the day but was very restless at night. In October he became worse and was sent in to Leith Hospital as a case of Chorea. While he was there his insomnia continued almost uninfluenced by treatment. He was very troublesome and impertinent owing/

owing to his habit of watching the other patients in the ward and making personal comments on their behaviour. He was abnormal, sharp-witted, and utterly lacking in shyness. Of the various drugs tried, the only one which had any effect was paraldehyde, which occasionally produced sleep. He was discharged from hospital in December 1920, his condition unchanged.

After leaving hospital he was at home for five weeks, without any improvement, and in the end of January 1921 he was admitted to the Sick Children's Hospital.

Immediately after admission he was quiet and slept well for four nights; he then became restless and did not sleep for more than one hour at night for nearly a week. At this time he was extremely bright and lively during the day, but pert and ill-mannered. Eventually sleep was induced by hot baths, and during the last fortnight of his stay in hospital he slept well at night. There was not, however, any corresponding improvement in his mental condition and he was still very unmanageable. He was discharged at the end of February, and went with his mother to Denmark, where he had a relapse, and was treated in the Children's Hospital in Copenhagen, where he improved slightly, but after discharge he became worse than he/



Case I. Showing Characteristic Attitude.

he had ever been. He returned to Leith at the end of September, and again improved for a time. In October his symptoms again became worse - he slept very little, and became naughty and disobedient. Towards the end of November he became definitely vicious for the first time. He bit his mother and sister if they attempted to interfere with him at night, and began to use bad language. Punishment had no effect: he got worse instead of better under reproof. By the beginning of December the boy had become almost maniacal, and quite unmanageable at home. He was brought back to the Children's Hospital and came under my observation.

State on Examination, 11th December, 1921.

A well grown but rather thin boy, with a curious blank expression and an inane smile. He stood stiffly, with his shoulders rounded and his arms hanging down in front of him. He also walked rather stiffly, but his gait was not particularly characteristic except in so far as it was to a certain extent reminiscent of paralysis agitans.

Thoracic and abdominal organs were healthy. He had a good appetite; the bowels were regular. Urine normal. The reflexes were normal: no Babinski; no Rombergism; no katatonia. He was restless and fidgetty and seemed incapable of standing still for a moment or two.

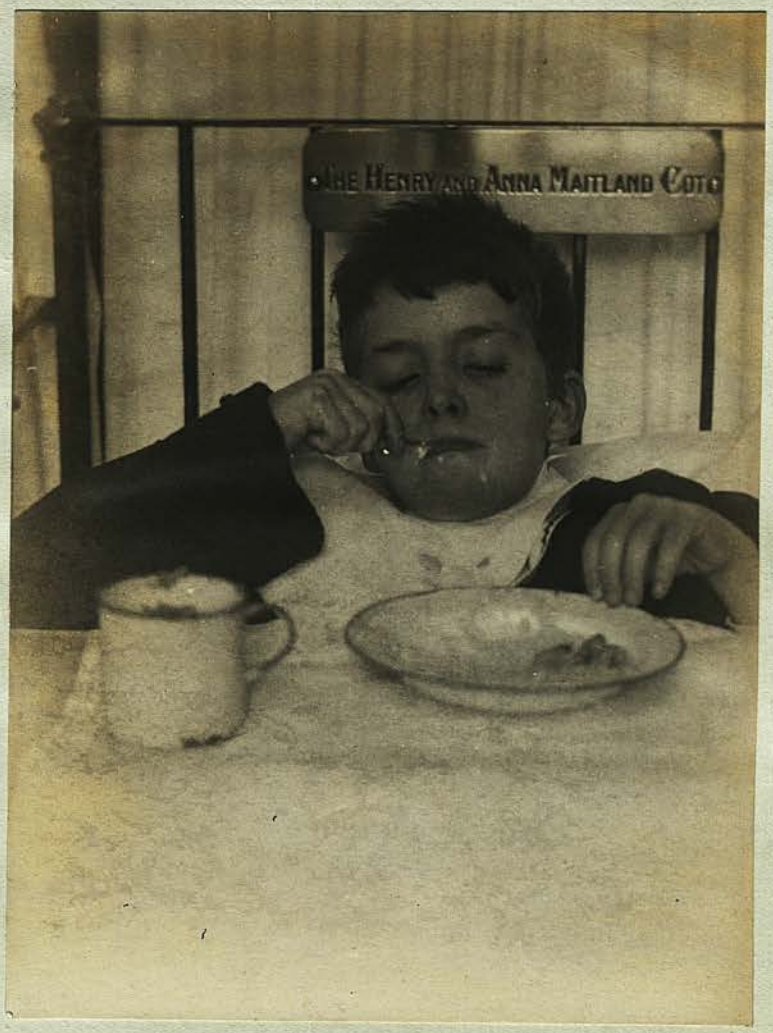
Mental Condition. The boy answered questions quite sensibly and seemed to have a good memory. He remembered having been in the ward 10 months before, and told various nurses that he knew them. Every five or ten minutes he had an attack of stertorous breathing in which he sat up in bed and puffed and snorted for several minutes, becoming very flushed the while; somewhat similar attacks of grunting occurred occasionally during sleep. He was uncleanly in his habits, and had a voracious appetite. He wolfed his food like a little animal; he constantly asked for more, and would have eaten several dinners in succession if he had been allowed to do so.

Examined by the Binet Simon Tests he was shown to have a mental age of 6½, his real age being 9¼: Intelligence Quotient = 81. He thus had a high intelligence quotient; he was good at tests involving knowledge acquired in the earlier years, but unsuccessful with comprehension tests, and was unable to adapt himself to novel problems. He was good at problems involving figures.

Progress. For three weeks after the child came under observation he was very troublesome. He shouted out at all hours of the day or night/

night, and if instant attention was not paid to him he banged on his locker. If any one came into the ward and did not speak to him he at once shouted to them to come and see him. He was never maniacal, but was noisy and talkative at night. He had no delusions or hallucinations. His sleep was much disturbed and very intermittent; sedatives had little effect until large doses of chloretone were given, whereupon he slept heavily for several days, and on waking was a good deal better. He was less restless during the day, did not break toys as he had done, and the fits of stertorous breathing became at the same time less frequent.

This improvement has continued. Peter is now comparatively well behaved; he is up and running about the ward. He is still very assertive and fond of attention, but is moderately polite. He is kindly to the smaller children, and likes playing with and nursing them. He has become cleanly in his habits; his attitude and movements are not so stiff, and attacks of stertor are now rare. He sleeps well, and only shouts out occasionally, and has gained weight.



Case II Lethargy and "Mask-Like" Facies

CASE II. J.O. male, aged 10 years.

History of Present Illness.

This boy took ill suddenly on 21st December 1920. He complained of headache, and became very drowsy, so that he went to sleep on the slightest provocation. The somnolent condition lasted without change until his admission to the Children's Hospital on 3rd January 1921. Previous to the onset of the present illness he had always been a normal, well behaved child.

On admission he was found to be very thin and wasted; he was extremely lethargic, and slept continuously day and night. The reflexes were normal; katatonia was fairly well marked; there was an internal strabismus of the left eye, but it was uncertain whether this had developed since the onset of his illness or had existed before. The cerebro-spinal fluid was clear, not under pressure, and showed neither an increase of the cells nor any abnormal constituent. Colloidal gold test negative.

The lethargy varied in degree a good deal; on some days it was impossible to arouse the patient, on others he could reply to questions in a sleepy fashion. Even when he was most deeply asleep, however, if the bed clothes were turned down he slowly pulled them/

them over him again as if feeling the cold. During the period of somnolence he seemed to have some idea of the passage of time; after he had been in Hospital about three weeks he was asked one day when he was more responsive than usual, how long he had been in the ward, and to this he gave an approximately correct reply. It was found difficult to feed the child as he was generally too sleepy to swallow properly. He remained obstinately constipated, and aperients had little effect.

He continued in this state until the 1st March, 1921, when he wakened up sufficiently to shake hands with the nurses and doctors. From that date he improved steadily. He began to eat well though slowly, falling asleep between mouthfuls. Towards the end of March he had gained a good deal of weight, and was able to be up and running about the ward. He was also brighter mentally, and was able to answer questions intelligently though slowly.

As recovery from the lethargic state took place his mentality seemed to undergo a change. From being good he became very disobedient and mischievous, used bad language, and gave way to violent fits of temper. A striking feature at this time was the development of a voracious appetite; he seized every opportunity which/

which presented itself of stuffing himself with food and was constantly pilfering from the other children's trays and the ward kitchen.

He was discharged in May 1921, very much improved, quite lively and energetic, and quite free from somnolence. After discharge he remained comparatively well for several months and returned to school. In September, however, he relapsed, and the school authorities reported to his mother that he was constantly falling asleep in school. He was therefore readmitted to Hospital in October 1921, when he first came under my observation.

State on Examination, 4th November 1921.

He was a well nourished and well developed lad. He was very drowsy and fell asleep while standing on the floor. His face was somewhat mask like, and he smiled slowly in rather a silly manner. His attitude was similar to that of the previous case: he stood stiffly, with the shoulders hunched up, keeping his arms hanging down straight in front of him. All his movements were slow and deliberate and he walked with a stoop. Apart from the general lethargy which characterized all his movements, and the mental alteration to be described, there was little to note on physical examination. The reflexes were normal, and the squint noted above was still present to a slight degree.

Mental Condition. Mentally, he seemed fairly intelligent and answered simple questions well though slowly. Speech was drawling and indistinct. The Binet-Simon tests gave the following result: Mental age, 8¾ years; Real age, 10 years; Intelligence quotient, .81. He responded well to tests involving facts learned in earlier years, but was unsuccessful with comprehension tests. Tests involving figures were well done. The principal mental change appears to be in the direction of an alteration of his morals and conduct. This change has set in since his illness began.

Progress. While in hospital he was always on the alert for opportunities to steal food. He used to go into the ward kitchen when nobody was looking and take bread and butter, or in fact any eatable he could lay hands on. The only result of scolding was an amiable smile, and an immediate repetition of the offence. At times he was rude in a sleepy sort of way, and he occasionally lapsed into outbursts of bad language. During the five weeks he was under observation, he made no real progress. He was abnormally somnolent during the greater part of that time, but varied a great deal from/

from day to day. At times he would sleep for 24 or 36 hours at a stretch; at other times he was more wakeful and would run about the ward all day, but even at his best he was very liable to fall asleep at unexpected times, such as while at meals, when dressing, or when at play. His body weight fluctuated a good deal for no obvious reason, as he always had an excellent appetite and was free from digestive disturbance.

From the time of his discharge till February 1922 he has continued in much the same condition. He was sent back to school, but, as before, fell asleep over his lessons. On one occasion he attracted a crowd in the street by falling asleep in the standing posture.

CASE III. J.M., female, age 7.

History of Present Illness.

In June 1920 it was first noticed that the girl began to breathe in a stertorous manner, and that she was very wakeful at night. She slept a little during the day, for perhaps an hour at a time thus getting about four or five hours sleep in the twenty-four hours. This insomnia has persisted until December 1921, the date of her admission to Hospital. Coincidently with the onset of her illness her disposition changed completely, and from being an ordinarily well behaved child, she has now become extremely naughty and unmanageable. About February 1921 she began to be destructive; she tore her clothes and bed clothes, threw dishes, etc. about, and smashed things wholesale. Since then she has steadily become more violent and difficult to control. She has become liable to outbursts of temper during which she shrieks loudly and weeps. In one of these fits she tears and breaks everything she can lay hands on; e.g., she would seize a loaf from the table and break it up into small pieces. Correction at these times only makes her worse. All the time her physical health and appetite have been good, but she has become thinner, and is thought not to have grown at all since the beginning of her illness.

State on Examination, 15th December 1921.

A thin, underdeveloped, extremely restless and fidgetty child. Her skin was dry and harsh, and there was little subcutaneous fat. Her lips were dry and cracked, and the tongue was furred. Physical examination was otherwise negative, but owing to her great restlessness it was difficult to test the reflexes satisfactorily. She breathed at times in a stertorous manner, which was more marked if she was plotting some mischief.

Mental Condition. Mentally, she was in a state of wild excitement, bordering on mania. She did not appear to be delirious, but understood all that was said to her and could talk and answer questions sensibly, but was incapable of any sustained attention. She was disobedient and naughty, and wanted her own way in everything; if thwarted in the slightest she flew into a violent fit of temper, spat at the person who had crossed her, and swore vigorously. Sometimes she had fits of tears, and threw anything handy at the person who had annoyed her.

Immediately after admission she asked for a drink of water, and at once threw the mug at the child in the next cot. On one occasion she slipped out of bed/

bed and ran round the ward hitting the babies on the head with a hair brush. She had quiet intervals at times, when she would sit and watch the smaller children being bathed, and would talk about her baby sister at home. During the week she was in hospital she seemed possessed of the demon of unrest. She spent most of her time getting out of bed and running up and down the ward. When put back to bed she seized all the clothes and dragged them from the top to the foot; no sooner was this done and the bed made in this way than she undid it, and started over again. She had a passion for cleaning and polishing, and spent the whole of one night scrubbing out a playground in the ward.

She slept very little. Immediately after admission she slept for a couple of hours, and then wakened up very restless. After an hour or two she fell asleep again for a short time, and then wakened up restless and noisy. This continued for 48 hours, when she got worse and screamed continuously for six hours one night, and at the end of that time, though quite exhausted, she did not sleep. For three nights and two days she did not sleep for more than five minutes at a stretch, although large doses of different sedatives were tried. Eventually she got six hours sleep/

sleep after large doses of chloretone, and after that though the sleep was always bad there was no repetition of the prolonged screaming.

As it proved impossible to manage her in a general ward, she was discharged a week after admission *in statu quo*.

CASE IV. J.M., male, aged 8 years and 4 months.

History of Present Illness.

In May 1920 the child came home from school one day and vomited; he then fell sound asleep, and remained sleeping for three weeks. After he recovered from this somnolence he became very sleepless and his disposition changed completely. He became disobedient and destructive.

At first he was restless and sleepless at night, tossing about the bed and throwing off the clothes. When he did fall asleep, he used to waken up about midnight, screaming, and at times apparently delirious. He talked a great deal in his sleep, and repeated lessons learned at school during the day. At this time he was abnormally drowsy during the day, and used to fall asleep at odd times irrespective of what he was doing, e.g., while in class. An effort was made to keep him awake and busy all day in order that he might sleep better at night, but it did not produce the wished for effect. He has gradually become worse, until at the time he came under observation, he was sleeping very little, either by day or night. About once every three weeks or so he has a fit of somnolence, and sleeps for two to three days at a time.

Latterly/

Latterly he has become more destructive, and has developed the habit of tearing up his blankets into strips. He is easily led and will steal at the instigation of other children. For some months before admission the boy breathed in a stertorous manner, especially at night. The curious snorting breathing also occurred when he was contemplating mischief, or actually engaged in it.

State on Examination, 7th January 1922.

A well grown and well nourished boy. The digestive, circulatory, and respiratory systems showed no abnormality. The knee jerks were exaggerated, but apart from this the nervous system was normal so far as physical examination went.

His face had a somewhat mask-like expression, and his smile was vacant. When standing his posture was rather stiff and unnatural, with a tendency to stoop; his gait was not in any way characteristic, and he did not stand with the shoulders hunched up as the other patients did. He had frequent attacks of stertorous grunting breathing, which appeared to be beyond his control. He was fidgetty under examination, and answered questions quickly and sensibly. He was quite devoid of the shyness natural to a child/

child coming into new surroundings, and was assertive and rather exalted. He enjoyed attention, and insisted on receiving it.

Mental Condition. The Binet-Simon tests showed that his intelligence was lower than some of the other cases. His mental age was 6½ years; his real age was 8½ years; intelligence quotient = 72. Like the other patients, he gave satisfactory replies to questions involving knowledge acquired before the onset of his illness, but did not score well in subjects that he would have learned later.

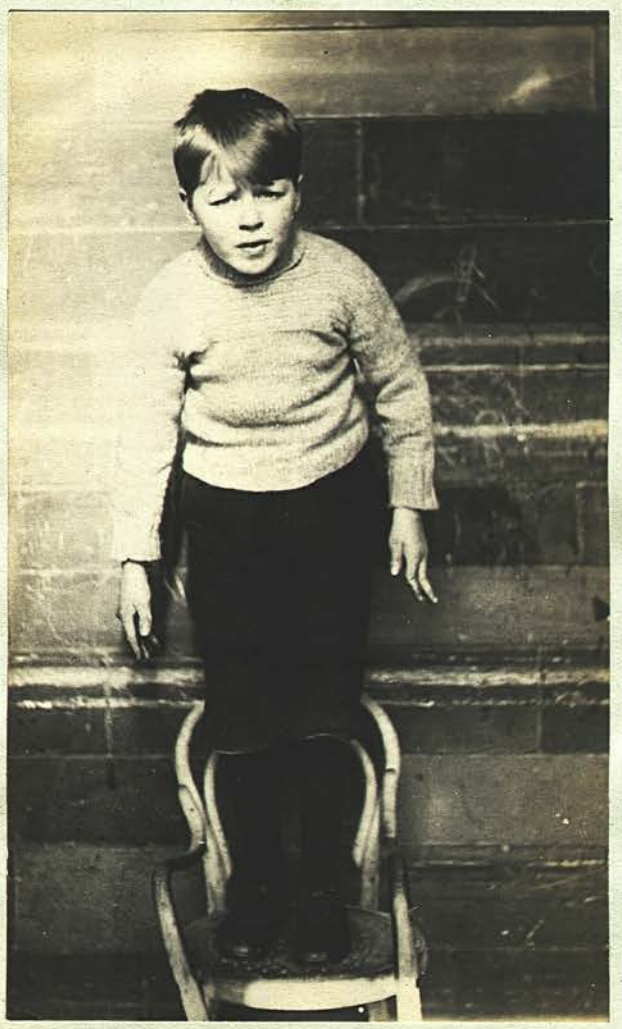
Progress. On his first two days in hospital he was fairly good, and slept the greater part of the time. He then became restless and slept badly, and began to tear up his blankets into strips, roll these up into a bundle and throw it on the floor. He gave no explanation of why he did this, except that he "liked it". Reproof had no effect in checking his destructiveness. He was in hospital for five weeks, and became progressively worse, so that ultimately he had to be sent home as unsuitable for a general ward. For a whole week he got practically no sleep at night, but sat up in bed arranging/

arranging the bed clothes, tearing up blankets and night clothes, and shouting out at intervals. Sometimes he shouted almost continuously during the whole day, but he was noisy in an aimless sort of way, and not violent and excited as is the case in acute mania. Even at his noisiest he would always stop and answer questions sensibly enough. Chloretone proved quite ineffective in his case; indeed, he seemed the worse of it. Luminal in doses of 1 grain twice daily soothed him to some extent, and he slept better while he was having it.

CASE V. H.H., male, aged 9 years and 3 months.

History of Present Illness.

In March 1918, during an epidemic of influenza, the boy had an attack of what was supposed to be that disease. He became very drowsy, and slept for eight weeks without waking, except occasionally for food. At the end of this time he began to recover, but had great muscular weakness for some months thereafter, and was unable to stand alone or to walk. Eventually he recovered sufficiently to return to school, which he continued to attend until the date of his admission to hospital in January of this year. Since September 1921, however, his condition has deteriorated; his left arm and leg have become weak and stiff, and his control of his movements has begun to fail, especially if he tries to do anything hurridly; if he is given plenty of time to execute a movement he does it pretty well, but he can do nothing abruptly. Accompanying this loss of control there has developed a degree of stiffness of the limbs and he is not able to use his hands properly. He walks with a curious stiff gait, "loping", in the father's phrase, and when once started off can only go/



Case V. Deformity of fingers and Attitude.

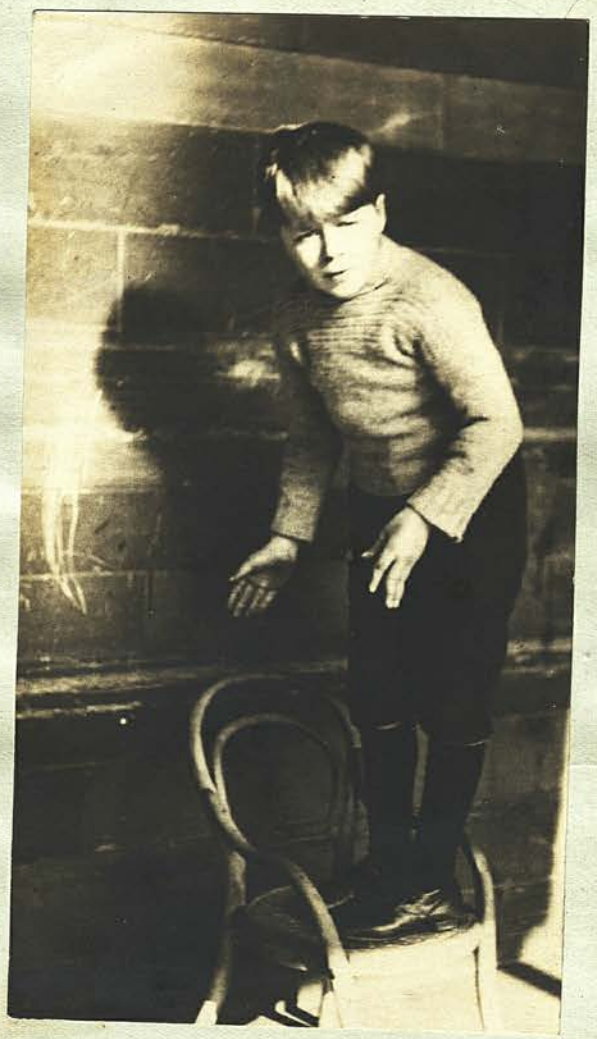
go in a straight line. He cannot turn quickly unless he stops himself by catching hold of something. If given plenty of time he can turn round.

He has slept throughout the course of his illness fairly well, though sometimes he is restless up till midnight, and he has once or twice walked in his sleep after any excitement in the day. He has a habit of sighing deeply at intervals, but there have never been any attacks of stertorous breathing.

Since the onset of his illness he has seemed dull and slow at grasping facts, but when once he has got a thing into his head he seems to retain it fairly well. His speech has become noticeably slow and indistinct.

State on Examination, 17th January 1922.

Well nourished and developed; appetite good; thoracic and abdominal organs healthy. He stands with a well-marked stoop, holding his hands stiffly in front of him with the arms partially extended. At first glance his appearance is suggestive of the hemiplegic type of spastic cerebral palsy, the left arm being held semi-flexed at the elbow, and the fingers hyperextended. The resemblance, however, is only superficial, for the apparent contractures can be completely overcome with very little application of/



Case V. Side view.

of force, and without causing him any discomfort. As soon as the arm is left alone it goes back slowly into its former position of flexion. The fingers of the left hand are hyperextended at the interphalangeal joints, and flexed at the metacarpo-phalangeal joints, and the thumb cannot be voluntarily opposed to the index finger distally to the first phalanx.
The result is that his grasp is awkward, and he is very clumsy in handling things and in feeding himself. The abnormal position of the hand can easily be remedied by pressure, but at once returns.
The right arm is affected in the same way, but to a much less degree. The legs also show a considerable amount of rigidity, especially on the left side.
Both knee jerks are sluggish; the plantar response is flexor on both sides, much brisker on the right than on the left. His gait is characterized by propulsion and retropulsion, resembling what is seen in paralysis agitans. He starts off in a straight line with the body stooped forward, and taking steps of a moderate length. As he lurches along his steps become shorter and shorter till he is trotting after his centre of gravity, as in a bad case of Parkinson's disease. When once he has attained a certain pace he cannot stop without catching hold of something, and/

and he often falls for this reason. At the same time, when his attention is fixed on his efforts, he seems to have the power of voluntarily walking with slow steps, and avoiding breaking into a run. He is apt to fall if told suddenly to turn round. When made to walk backwards he shows marked retropulsion, and trips over himself and falls if not caught. From time to time he gives a deep involuntary sigh.

Mental Condition. Mentally, he is emotional, and laughs and cries with little provocation. He has a rather mask like expressionless face, with the mouth held slightly open. He is dull and rather stupid, but probably he gives the impression of being more stupid than is actually the case on account of his slowness in answering and the expressionless face. He has a slow foolish smile. He is good-natured on the whole, and very inquisitive; always wishing to see what is going on when patients are being examined. Occasionally he is noisy and shouts a great deal. His speech is slow and indistinct, and his reaction time to questions is definitely delayed.

Examination by the Binet-Simon tests showed that his intelligence was low, his mental age being 6½/

6½ years as against his real age of 9¾; Intelligence quotient = .68. Like the other cases he gave satisfactory answers to questions involving knowledge acquired before the onset of his illness, but scored little on subjects he should have acquired later. He showed marked stereotypy, and the examination suggested that he was definitely feeble minded.

He is somewhat dirty and untidy in his habits and dress.

Progress. During the six weeks he has been under observation in hospital he has made no progress. When he is up he is inclined to be rather noisy, and is easily upset by the teasing of the other children.

CASE VI. G.P., male, aged 7 years.

History of Present Illness.

He took ill suddenly in January 1921; before this he was a bright intelligent child. The first symptoms noticed were twitchings of his arms and legs, with definite loss of power in the latter. Along with this his mental condition underwent a change. He became stupid, bad tempered, and emotional, laughing and crying much more readily than before. His speech also became affected about the same time; it became indistinct, and he spoke (in his mother's phrase) "as if his mouth was full of water". He became restless and sleepless at night; after being put to bed at the usual time he was in the habit of getting up and playing with some soldiers he possessed, and also of talking to a photograph of his dead father and asking it for pennies. He also seems to have been hallucinated at times; he said there was a hen in his bed, and started to hunt for it. Towards morning he used to fall into a peaceful sleep which would last into the forenoon, and as the day wore on he would brighten up again and become active towards nightfall. This state of matters/

matters continued for a month; he was then put to bed altogether, and kept there for six months. During this time he improved considerably in every way, but as soon as he got up he relapsed, and was soon as bad as ever, being very nervous and excitable.

At the beginning of November 1921 he began to drag his left leg as he walked, and could only walk for a short distance. This continued until he came under observation in January 1922.

State on Examination, 10th January 1922.

The child is pale and badly nourished. The skin is rather dry. The teeth are carious; the tongue is furred and shows a coarse tremor. Apart from the nervous system there is no noteworthy abnormality of any of the organs.

His gait is very unsteady, and can best be described as spastic, with a clumsy staggering ataxic character. He has difficulty in controlling the left leg, which is hyperextended at the knee when he takes a step. He walks with considerable effort and difficulty.

He shows a definite spastic condition of the left side, with exaggeration of the reflexes. There is no Rombergism, and the sense of position is good: no true ataxia. Both knee jerks are increased;

knee/

knee clonus is present on both sides, and ankle clonus on the left side. The plantar response is flexor on the right, extensor on the left side. There is slight wasting of the muscles of the left leg; the muscles are somewhat rigid, and there is distinct loss of power. There is no tremor or other involuntary movement.

Mental Condition. Mentally, he is very good natured, talkative, and self-assertive. He is very impatient if his wishes as regards food, toys, etc. are not at once attended to. His conversation is foolish, but he is sharp enough in some ways, quickly picking up any remark that refers to himself; his memory is good. He cannot read, but can count fairly well. His speech is slurred and indistinct.

Throughout the day he is quiet and well behaved, but towards night he rouses up and begins to get lively. He will sleep for an hour or so, and then waken up and play with his soldiers. He then falls off again for an hour, and repeats the programme over again. He has no Hallucinations. His condition has not changed while in hospital.

CASE VII. J.B., female, aged 3¼ years.

History of Present Illness.

In February 1919, when the child was 10 weeks old, she took a fit of crying which lasted all night. Next day her eyes were seen to be staring, and she did not seem properly conscious, and during the following night she took a fit of short duration, which recurred at intervals until the morning. She was treated with bromide, and had no more fits. After this she seemed to sleep day and night for a week, and paid no attention to anything. She took her food well during this time, but in spite of this she wasted a good deal. After her illness had lasted a week she was admitted to the Sick Children's Hospital on 6th February 1919. On admission she was found to be a well nourished child, who was constantly jerking her legs in a purposeless spastic manner. There was some nystagmus, and slight head retraction. The reflexes were increased, and there was double Kernig. Otherwise nothing was made out, and the case was diagnosed as meningitis.
The results of lumbar puncture, however, were peculiar, and did not substantiate this diagnosis. She was punctured five times on successive days, and on each occasion/

occasion nothing but pure blood was withdrawn; thereafter several more spinal punctures were done, and the fluid was found to become gradually less blood stained and finally quite clear and apparently normal. No growth was ever obtained from the fluid. She was discharged from hospital at the end of five weeks, somewhat improved. Her head symptoms were less marked, and she was brighter than on admission, but had lost a considerable amount of weight. No definite diagnosis was made at that time.

After leaving hospital she thrived well, but made little or no progress mentally. She was re-admitted to hospital on account of a gastro-intestinal upset on 29th October 1919. At that time she was a heavy, stupid looking child who opened her eyes with difficulty. She had double ptosis, contracted pupils, and vertical and lateral nystagmus. She could, however, follow moving objects. The fundi were normal. She held up her head fairly well, and could sit up with support, but had made no attempt to speak or walk, and never cried. She had occasional slow twitching of her face; no fits.

The knee jerks were present; left less than right. No head retraction. Double Kernig, best marked on right. Abdominal reflexes absent. She slept/

slept well at night. The cerebrospinal fluid was normal. An X-ray of the cranium revealed nothing to account for her condition. She was discharged from hospital after five weeks residence in the same condition as on admission.

After her return home she made slow physical and mental progress. Up to the age of two years she took little notice of anything. About that time she began to grasp things in her hands and look about her; she began to distinguish her father from her mother, and strangers from either. She can now distinguish between different kinds of food, and seems to understand a little of what is said to her, but cannot be trained in any way. She cannot stand or feed herself, and is incontinent. During this period of her illness her mother noticed that the child had the habit of giving frequent deep sighs.

In October 1920, one year and eight months after the beginning of her illness, her sleep became much disturbed. She is restless after she is put to bed at 7 o'clock, and lies kicking the bedclothes about. About midnight she begins to scream, and goes on until she is exhausted. She falls asleep about 4 a.m., and sleeps soundly till mid-day, being difficult to rouse during this time. This state of matters lasts for/

for a month or so and then there is a remission during which she sleeps quite well without any screaming. Her mother has brought her up to hospital at intervals on account of her extreme restlessness and sleeplessness, and various sedatives have been employed, but without avail.

State on Examination, 30th January, 1922.

The child is heavy looking, and obviously mentally defective; she pays no attention to anything that is said to her. She is very restless, constantly throwing her limbs about. This, the mother states, continues during sleep. From time to time she gives a very long deep sighing respiration. She cannot stand by herself, but can take a few staggering steps if firmly supported. The abdominal and thoracic organs appear normal.

Nervous system. Knee jerks present; sluggish.
Double flexor response. No Kernig.
Abdominal reflexes present.
Pupils normal.

Mental Condition. Mentally the child seems very retarded, and understands very little of what is said to her. Asked where her mother was she turned her eyes in that direction, but made no attempt to point to her. Bright objects attracted her, and she tried to grasp at them.
She was not admitted to hospital, as at present she is sleeping fairly well.

COMMENTARY. The seven cases recorded above illustrate different types of the disease, in which the leading features are:- extreme lethargy (Case II), insomnia and restlessness (Cases I and IV), great mental excitement (Case III), the "paralysis agitans" syndrome (Case V), true spastic paralysis (Case VI), and marked mental deficiency (Case VI).

ONSET. There are two chief modes of onset. In one, which approximates to the adult type of the disease, there is extreme lethargy. This lasts for several weeks, during which the patient sleeps more or less continuously, and is roused with difficulty for food, and to evacuate the bladder and bowels. Such cases are described by Batten and Still, and Finlay in their early papers. They may or may not develop insomnia later on.

In the other form of the disease there is no history of a period of lethargy. The child suddenly ceases to sleep at night, becomes restless and very wide awake towards evening, continues thus till the early hours of the morning, and then falls asleep. This sleep lasts well into the forenoon, and once asleep, the child is difficult to rouse. Many of these/

these cases have also choreiform movements. Finlay and Shiskin find these two symptoms the most outstanding in their series of cases.

SYMPTOMS AND SIGNS. As might be expected from the manifold symptomatology of lethargic encephalitis as seen in the adult, even in this small series of cases the signs display considerable variation, and it is not suggested that they by any means cover all the possible changes. Certain phenomena, however, appear to be fairly constant, and are therefore of greater diagnostic importance.

In addition to the characteristic sleep disturbance referred to, the peculiarities of respiration, the attitude and gait, the mask-like facies, and the alteration in the mental condition may be noted from this point of view.

The children are, as a rule, well nourished, and show no abnormal signs or symptoms in the thoracic or abdominal organs.

REFLEXES. The reflexes vary considerably, in some cases they are unaffected, in others they show considerable disturbance, varying from increased or diminished Knee-jerks, to marked/

marked changes, such as a positive Babinski, which accompany cases with a true upper neuron paralysis.

ATTITUDE AND GAIT. The attitude of these children is characteristic. The child stands in a stiff, unnatural manner, with his shoulders hunched up, his head poked forward and his hands hanging down in front of him. If told to hold himself straight, he can do so, but as soon as his attention is relaxed, he returns to his old attitude. In one case, (V), this occurs to such a marked extent that the child is apparently suffering from spastic paralysis. This is not so, however, as all the "contractures" can be easily overcome. This condition, which appears as a late symptom, is probably related to the catatonia which occurs as an acute symptom (Case II), and is described by Batten & Still, and Finlay in their early papers.

The gait of these children is stiff and unnatural. The child may take short steps, which tend to become shorter and quicker as he proceeds. In the very marked cases, the gait develops into regular propulsion and retropulsion (Case V), and consequently the child has some difficulty in moving about.

FACIES./

FACIES. The facies in these cases is usually very expressionless, "mask like". There is an absence of wrinkles, and a flattening of the naso-labial fold. Emotion produces very little change of expression, and when the child is amused the result is a silly smile. Consequently the child gives a rather erroneous impression of marked mental defect.

BREATHING. A fairly constant feature of the disease in these cases is a change in the character of the respirations. In the more marked cases the child sits up in bed, and begins to take long, deep, breaths, with a sighing expiration. These breaths become more rapid and noisy, and finally the child is puffing and blowing and snorting in a most extraordinary fashion. During the attack the child becomes very flushed. The attack lasts for 3 - 5 minutes, and then passes off, only to be repeated at a short interval of time. It is quite involuntary, and also occurs during sleep. In the less marked cases, the child gives a frequent deep involuntary sigh.

The above respiratory rhythm is a very striking symptom, which has hitherto escaped the notice of most/

most writers. The only reference to it in the literature is by Fletcher and Rolleston, who describe a case in which it was present. Rolleston comments on this case, and points out the probable connection between this phenomenon, and the epidemic hiccough, yawning, etc. which is connected with epidemic encephalitis.

In the majority of cases the general physical picture presented by the children is reminiscent of paralysis agitans, except that none of them have tremors. Tremors closely resembling those of paralysis agitans have, however, been recorded by Paterson and Spence, as occurring along with other symptoms such as stiffness, chiefly as a late manifestation of the disease.

MENTAL EFFECTS. In the early stages of the disease, the mental condition of the child depends on the mode of onset. In the lethargic cases there is a general delay in comprehension, response, etc., corresponding to the degree of lethargy. Even the very marked cases, however, are not really comatose, and have a definite knowledge of the passage of time (Case II). This fact was noted also by Batten & Still in their early cases.

In/

In the cases in which the onset is marked by insomnia, the child is usually abnormally alert mentally, especially at night. He is not delirious, but is very talkative, and notices, and comments on, little happenings which a normal child apparently does not observe.

The later mental effects of the disease are much the same in all cases.

The child's intelligence is not markedly impaired, but seems to stop short at the point at which his illness commenced. Consequently, the earlier in life the child develops the disease, the more deficient in intelligence he appears later, and children who acquire the disease in infancy are often very low grade imbeciles (Case VII). Paterson & Spence have also observed this.

The results of the Binet-Simon tests demonstrate this same fact. Children who have had the disease for a long period have a lower mental age and intelligence quotient than others of the same age who have been ill for a shorter period (Case I and Case V).

The most noticeable mental change, however, is one of character, conduct and morals. The child, who up to the time of his illness was normal, and moderately well-behaved, suddenly changes completely.

He/

He becomes very naughty and unmanageable. He is emotional, and seems to have lost his self-control. He laughs and cries much more easily than formerly, and is subject to violent outbursts of temper. He is frequently impertinent and disobedient, and uses bad language. Reproof and punishment have no effect upon him. He is utterly lacking in shyness, is self-assertive, and extremely egotistical.

He is childish for his age, untidy in dress, and dirty in habits. He is often destructive in an aimless fashion, tears things up, and smashes them, but does not know why he does it.

He has an abnormal appetite, will eat all day long if permitted and will steal food to satisfy his greed.

In looking round for a comparison, the mental disturbance which in a general way presents analogies with this one is General Paralysis of the Insane in its early stages, and it may be noted that in respect of their infective origin, the two diseases may be compared with one another. This similarity is also commented on by Leahy and Sands.

SEX INCIDENCE. The sex incidence is interesting, inasmuch that more males than females seem to suffer from the disease.

In/

In my own series of cases there are 5 boys and 2 girls. Of the 24 cases recorded by Finlay & Shiskin 19 boys and 5 girls suffered from the disease. Happ and Blackfan in a series of 6 cases had 5 boys and 1 girl. Leahy and Sands series include 4 boys and 2 girls. Happ and Mason, however, have equal numbers of each sex in the 24 cases recorded as occurring in children. Paterson and Spence do not mention the sex of their 24 cases.

COURSE. The course of the disease is much more chronic than was at first suspected. Many cases which were apparently cured have had relapses, or have developed late symptoms months, or even years after the onset of the disease, while others have slowly and steadily deteriorated from the beginning.

This is borne out by my own series of cases, none of which have really recovered, although suffering from encephalitis for months, or even years. Paterson and Spence lay special emphasis on this fact, and record cases of relapses in children who have previously been reported cured. Happ and Blackfan also remark on the extreme chronicity of the disease.

PROGNOSIS./

PROGNOSIS. The disease has been known for too short a time to allow of any dogmatic statement as to prognosis, but in many cases it does not appear to be very favourable, especially if they are of long duration, although a considerable number of recoveries are recorded.

TREATMENT. Treatment in these cases is very unsatisfactory. Many cases improve considerably when treated for long periods in an institution but relapse, and become utterly unmanageable, if sent home. Leahy and Sands have apparently had similar results with their cases.

Insomnia is very difficult to treat. Warm baths at night sometimes help. Leahy and Sands recommend warm packs as an alternative. Sedatives are of little use, although sometimes large doses of chloretone or luminal produce the desired effect. Efforts to tire the child out during the day seem worse than useless. Many children sleep better after a time, as a result of the general improvement produced by institutional treatment.

Stiffness and paralysis are very intractable, and massage has little effect.

Mental deterioration, and gross moral defects such/

such as persistent stealing, are difficult to do anything for, as punishment has no effect, and the child has no sense of shame.

Treatment, on the whole, is therefore disappointing and the only possible chance of any improvement seems to lie in prolonged residence in an institution.

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